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Two-Dimensional Echocardiography in the Pre- and Postoperative Management of Totally Anomalous Pulmonary Venous Connection

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The records of 23 infants who underwent surgical repair of isolated totally anomalous pulmonary venous connection were reviewed to assess the accuracy of pre- and postoperative echocardiographic diagnoses. Preoperative echocardiographic diagnoses were accurate in 22 of 23 patients, including the sites of connection of the individual pulmonary veins. Cardiac catheterization in 13 patients confirmed the echocardiographic findings. Analysis of multiple pre- and postoperative variables revealed no statistically significant difference between the infants with and without catheterization, although there was a tendency toward a higher mortality rate in the catheterized group.

Postoperative echocardiographic examination revealed obstruction to pulmonary venous return in 7 of 19 patients. Catheterization confirmed the echocardiographic findings, localizing the obstruction in one patient.

The size of the venoatrial anastomosis was measured on postoperative echocardiograms performed on 14 patients. The cross-sectional area of the anastomosis was <0.3 cm^2/m^2 of body surface area in the four patients with obstruction of the anastomosis, and >0.95 cm^2/m^2 in all long-term survivors examined.

Two-dimensional echocardiography with pulsed Doppler examination and Doppler color flow mapping is an excellent means of diagnosing totally anomalous pulmonary venous connection. The connections of the individual pulmonary veins can be identified in nearly all cases. Surgical repair can usually be undertaken on the basis of echocardiographic diagnosis alone. Echocardiography also provides an extremely accurate method of evaluating surgical repair and of identifying and localizing postoperative obstruction to pulmonary venous return.

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Optimal surgical correction of totally anomalous pulmonary venous connection requires accurate and complete delineation of the anatomic details. Two-dimensional echocardiography offers an excellent noninvasive means of diagnosing totally anomalous pulmonary venous connection (1–5). In addition, Doppler color flow mapping facilitates identification of the precise site of connection of each individual pulmonary vein.

Recently, repair of totally anomalous pulmonary venous connection diagnosed by echocardiography, without preoperative cardiac catheterization, has been reported (6,7). Since the diagnosis is usually made in the neonatal period, often in critically ill infants for whom invasive procedures carry increased risk (8), rapid noninvasive diagnosis is desirable. In late 1984, we began operating on selected neonates with totally anomalous pulmonary venous connection without cardiac catheterization if each of the individual pulmonary veins was visualized echocardiographically.

A retrospective study was designed to assess the accuracy of two-dimensional echocardiography in identifying the precise anatomic details in totally anomalous pulmonary venous connection, and to determine what additional information is obtained by cardiac catheterization. The role of echocardiography in the postoperative management of totally anomalous pulmonary venous connection and pulmonary venous obstruction was also examined, and a method was devised for assessing the size of the venoatrial anastomosis.

Methods

Study patients. All infants who underwent repair of isolated totally anomalous pulmonary venous connection at The Children's Hospital in Boston between January 1983 and January 1989 and who underwent two-dimensional echocardiographic examination at this hospital were identified by a search of the computerized cardiology data base. Patients with heterotaxy syndrome and other associated congenital cardiac anomalies were excluded. Each patient's medical records, echocardiographic reports, catheterization report,

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operative notes and long-term follow-up information were reviewed.

Preoperative echocardiographic evaluation. Echocardiographic examinations were performed with either a Hewlett-Packard model 77020 or an ATL Mark 600 cardiac imager, using a 5 MHz transducer. Chloral hydrate, 60 mg/kg body weight orally, or morphine sulfate, 0.05 to 0.1 mg/kg intravenously, was used for sedation when necessary. Subxiphoid, apical, parasternal and suprasternal notch views were used in all patients. Doppler color flow mapping was employed in the nine infants who presented after December 1986. The diagnosis of totally anomalous pulmonary venous connection was made by using criteria described previously (4,5).

The individual pulmonary veins and their site or sites of connection were identified and examined for signs of obstruction when possible (5,9).

Operative management. All infants underwent surgical repair using cardiopulmonary bypass and deep hypothermic circulatory arrest. Operative findings were compared with preoperative echocardiographic diagnoses.

Impact of cardiac catheterization. Infants who did not undergo preoperative cardiac catheterization (Group 1) were compared with those who did (Group 2) with respect to age at presentation, preoperative clinical status and type of pulmonary venous connection. Age at operation, date of surgery and cardiopulmonary bypass and deep hypothermic circulatory arrest times were also compared. Postoperatively, duration of mechanical ventilation, intensive care and hospital stay as well as late mortality and symptoms at follow-up study were compared.

Postoperative evaluation. Pulmonary venous obstruction was suspected postoperatively if one or more of the following echocardiographic findings were present: 1) A small anastomosis between the pulmonary venous confluence and the left atrium; 2) a continuous, nonphasic, relatively high velocity Doppler flow signal at the level of the anastomosis or pulmonary vein orifices; and 3) systolic flattening of the ventricular septum suggesting pulmonary hypertension.

Anastomosis size. The anastomosis between the pulmonary venous confluence and the left atrium was measured when possible on subxiphoid long- and short-axis images (Fig. 1 and 2). The cross-sectional area of the anastomosis was estimated by using the formula for the area of an ellipse (1/2 long-axis dimension × 1/2 short-axis dimension × π), and was subsequently indexed for body surface area.

Statistical analysis. Wilcoxon rank sum and Fisher exact tests were employed to test the significance of differences between Group 1 and Group 2.

Results

Patient characteristics. The 23 patients studied are described in Table 1. Four other patients underwent repair of totally anomalous pulmonary venous connection during the study period but did not undergo two-dimensional echocardiographic examination at this institution.

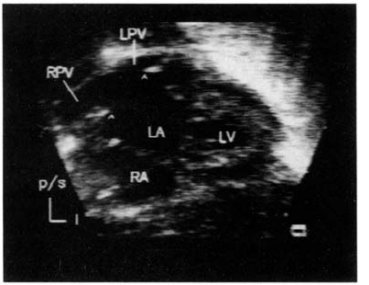

Figure 1. Subxiphoid long-axis view of the anastomosis between the pulmonary venous confluence and the left atrium (LA). **Arrowheads** ( ) indicate margins of the anastomosis. l = patient's left; LPV = left pulmonary veins; LV = left ventricle; p/s = posterior and superior (cephalad); RA = right atrium; RPV = right pulmonary veins.

Age at presentation ranged from 1 to 98 days (median 7). Thirteen of the 23 infants underwent preoperative cardiac catheterization, 4 at the referring hospital. Of the nine catheterizations at this institution, four were performed before December 1984, after which we began operative repair in selected infants without cardiac catheterization. Five patients underwent catheterization between December

Figure 2. Subxiphoid short-axis view of the anastomosis between the pulmonary venous confluence (PV conf) and the left atrium (LA). **Arrowheads** (^) indicate the margins of the anastomosis. a = anterior; desc Ao = descending aorta; RA = right atrium; RPA = right pulmonary artery; s = superior (cephalad).

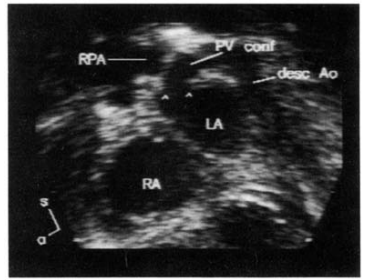

Table 1. Description of Findings and Outcome in 23 Patients

Pt No.	Age at Presentation (days)	Echo Diagnosis	Documentation of Obstruction	Outcome	Follow-Up (mo)	Anastomosis Size (cm^2/m^2)
			Group 1 (with cardiac catheterization)			
1	0	Subdiaph.	Echo	Asymptomatic	49	1.18
2	22	Subdiaph.		Asymptomatic	6	
3	21	Subdiaph.	Echo	Asymptomatic	4	1.38
4	28	Subdiaph.	Echo	Asymptomatic	5	
5	26	Subdiaph.	Echo	Asymptomatic	24	1.53
6	2	To coronary sinus		Asymptomatic	36	0.98
7	98	To coronary sinus		Asymptomatic	14	
8	3	Supra. to L. innom. v.		Deceased	4	1.50
9	1	Supra. to L. innom. v.		Asymptomatic	19	
10	0	Mixed	Echo	Asymptomatic	35	0.96
			Group 2 (without cardiac catheterization)			
11	0	Subdiaph.	Cath	Asymptomatic	63	
12	0	Subdiaph.	Echo, Cath	Deceased	6	0.24
13	1	To coronary sinus		Asymptomatic	6	
14	0	To coronary sinus	Echo, Cath	Deceased	4	0.25
15	30	To coronary sinus	Cath	Asymptomatic	43	2.16
16	0	Supra. to L. innom. v		Recurrent pneumonia	70	
17	7	Supra. to L. innom. v.		Deceased (liver disase)	12	0.96
18	95	Supra. to L. innom. v.	Echo	Asymptomatic	20	
19	84	Supra. to L. innom. v.		Asymptomatic	33	
20	88	Supra. to RSVC	Cath	Asymptomatic	10	2.27
21	28	Supra. to RSVC/RA		Asymptomatic	48	1.71
22	49	Supra. to RSVC*	Echo, Cath	Deceased	2	0.23
23	0	Mixed	Echo, Cath	Deceased	6	0.28

*Supracardiac anomalous pulmonary venous connection to the right superior vena cava by echocardiography and angiography. At initial operation, only a subdiaphragmatic connection was found; reoperation revealed an additional connection to the right superior vena cava. Cath = cardiac catheterization; Echo = echocardiography; L. innom. v. = left innominate vein; Mixed = mixed pulmonary venous connections; RSVC = right superior vena cava; RSVC/RA = junction of right superior vena cava with right atrium; Subdiaph. = subdiaphragmatic pulmonary venous connection; Supra. = supracardiac anomalous pulmonary venous connection.

1984 and January 1989 at the request of the individual cardiologist or surgeon caring for them.

Preoperative echocardiographic evaluation. Echocardiography correctly diagnosed the site of all pulmonary venous connections in 22 of 23 patients. The site was subdiaphragmatic in seven patients, to the coronary sinus in five and supracardiac in eight. Two patients were correctly diagnosed as having multiple sites of pulmonary to systemic venous connection ("mixed" totally anomalous pulmonary venous connection). Patient 10 had connection of the venous confluence both to a left ascending vertical vein and to a descending subdiaphragmatic vein. In Patient 23, the right pulmonary veins and the left lower vein connected to the coronary sinus, whereas the left upper vein connected to the left innominate vein, with a small additional connection to the coronary sinus.

The echocardiographic diagnosis was incomplete in a third patient with "mixed" totally anomalous pulmonary venous connection. This patient (Case 22) will be discussed later under "Operative Management."

Although diagnosis may have been facilitated by Doppler color flow mapping, no statistically significant difference in accuracy of diagnosis was found when this method was used.

Various degrees of preoperative obstruction to pulmonary venous return were identified by echocardiography or catheterization, or both, in 13 of the 23 patients (Table 1).

In the 23 patients presenting during the study period, all diagnosed by echocardiography as having isolated totally anomalous pulmonary venous connection, no significant additional cardiac lesion was identified by catheterization (if performed), surgical findings or subsequent follow-up study.

Operative management. In 22 of the 23 infants, the diagnoses at operation were the same as described by echocardiography. Surgical exploration revealed an additional finding in one patient (Case 22) with mixed totally anomalous pulmonary venous connection. This patient was diagnosed by echocardiography and angiography to have a small, tortuous vein connecting the pulmonary venous confluence to the superior vena cava. This connection was not found at initial operation. Instead, a small descending vertical vein coursing below the diaphragm was divided. Because of subsequent clinical deterioration with evidence of a persistent left to right shunt, the patient underwent reoperation,

when the connection to the superior vena cava was identified and ligated.

Impact of cardiac catheterization. All nine patients who underwent cardiac catheterization at this institution had an echocardiographic diagnosis of totally anomalous pulmonary venous connection before catheterization. The four patients who underwent catheterization at referring hospitals underwent echocardiographic examination at Children's Hospital after transfer.

In all 13 infants who underwent cardiac catheterization, the anatomic diagnosis was the same as that described by echocardiography at this institution. In Patient 22, catheterization failed to demonstrate the additional subdiaphragmatic connection of the pulmonary veins subsequently found at operation. No patient had any apparent morbidity as a result of catheterization.

There was no difference between Group 1 (without catheterization) and Group 2 (with catheterization) with regard to age at presentation ($p > 0.05$), or need for mechanical ventilation ($p > 0.5$) or intensive care ($p > 0.1$) preoperatively.

There was no significant difference in age at operation between Group 1 (mean 30 days, range 1 to 112) and Group 2 (mean 39 days, range 2 to 98). There was also no significant difference in date of surgical repair or in duration of totally cardiopulmonary bypass, circulatory arrest or aortic cross-clamp time during surgical repair ($p > 0.05$). Finally, there was no difference in duration of ventilatory support, intensive care or hospitalization after surgery.

Postoperative evaluation. There were no deaths in the immediate postoperative period or during the same hospitalization. Five infants eventually died from pulmonary venous obstruction (one patient in Group 1 and four patients in Group 2). A sixth patient died of biliary atresia at 1 year of age. Survivors were followed up from 4 to 70 months postoperatively.

Echocardiographic evaluation of postoperative venoatrial obstruction. Nineteen patients underwent postoperative echocardiographic examination. Twelve patients had no evidence of obstruction by echocardiography. Except for the patient with biliary atresia, all of these patients are alive and asymptomatic at long-term follow-up.

Seven patients had postoperative echocardiographic evidence of obstruction to pulmonary venous return by imaging or Doppler ultrasound, or both. Five of the seven patients were symptomatic, with onset of symptoms between 15 days and 2 months after initial surgical repair. Obstruction appeared to be at the venoatrial anastomosis in four of the five symptomatic patients (Cases 12, 14, 22 and 23). The remaining symptomatic patient (Case 8) had right ventricular hypertension by two-dimensional echocardiography, but the site of obstruction could not be identified echocardiographically. This patient is discussed later in this section.

Although all four patients with obstruction at or near the site of anastomosis underwent surgical revision of the venoatrial anastomosis, all four died of pulmonary venous obstruction between 1 and 5 months after reoperation. Obstruction was demonstrated by serial echocardiograms to be progressive in three of the four patients. The fourth patient had progressive obstruction demonstrated before reoperation but did not undergo repeat echocardiographic examination in the 4 weeks between reoperation and death.

Obstruction was found at one (Case 10) or multiple (Case 16) individual pulmonary vein orifices in the two asymptomatic patients. Patient 10 did not undergo reoperation and is asymptomatic at latest follow-up. Patient 16 underwent surgical exploration of the individual pulmonary veins. The left pulmonary veins could not be found; the anastomosis between the right pulmonary veins and the left atrium was revised. This patient, who has Treacher-Collins syndrome, a repaired tracheoesophageal fistula and a tracheostomy, continues to have occasional respiratory infections and receives long-term diuretic therapy. Subsequent echocardiograms have not demonstrated any obstruction to the remaining pulmonary veins.

Impact of postoperative catheterization. All seven patients with echocardiographic evidence of pulmonary venous obstruction underwent cardiac catheterization. In six patients, the predicted site of obstruction was confirmed. Patient 8, in whom the site of obstruction could not be localized echocardiographically, was found at catheterization to have inoperable long segment stenosis of all of the individual pulmonary veins. Cardiac catheterization was instrumental in identifying the site of obstruction in this patient, who died without undergoing reoperation.

Three asymptomatic patients, with no evidence of obstruction by postoperative echocardiography, underwent postoperative cardiac catheterization. The findings at catheterization did not differ significantly from the echocardiographic findings. Two patients underwent neither echocardiography nor catheterization in the follow-up period, and are clinically well.

Anastomosis size. Postoperative echocardiograms were adequate for measurement of the venoatrial anastomosis in 14 patients. The cross-sectional area of the anastomosis was calculated (in three before reoperation and in one after reoperation) as <0.3 cm^2/m^2 in all four patients who eventually died of obstruction of the anastomosis. In contrast, the cross-sectional area of the anastomosis was greater than 0.95 cm^2/m^2 for all 10 survivors whose anastomosis could be measured retrospectively. Patient 8, with stenosis of all of the individual pulmonary veins, had a large anastomosis (1.5 cm^2/m^2).

Discussion

Preoperative evaluation. Several investigators (1,3,6,7, 10–13) have reported their experience repairing various types of congenital heart defects without preoperative cardiac catheterization. To justify this approach, certain criteria must be met (3). The echocardiographer must be confident that the cardiac anatomy has been seen well and that the

correct diagnosis has been made; and the surgeon must have confidence in the echocardiographic findings and must be cognizant of the limitations of the technique.

If the echocardiographic diagnosis is incomplete, catheterization should be performed. Imaging pulmonary veins beyond their confluence is impossible in some patients, making it difficult to rule out distal obstruction. If the echocardiographic findings are inconsistent with the clinical presentation or course, confirmation of the diagnosis by catheterization is warranted.

It has been well documented that the diagnosis of totally anomalous pulmonary venous connection can be made noninvasively. Our data further indicate that echocardiography with pulsed Doppler ultrasound allows identification of the site or sites of connection of the individual pulmonary veins with such accuracy that catheterization is unnecessary in most cases. Given the high degree of accuracy of echocardiographic diagnosis in the study patients, the additional benefit provided by Doppler color flow mapping is difficult to quantify; however, this technique appears to facilitate localization of the connections of the individual pulmonary veins, especially when multiple sites of connection are present. Alterations in the Doppler color flow map suggesting turbulence or elevated velocity simplify the identification and localization of obstruction to pulmonary venous return.

When obstruction is suspected, surgical repair should be performed promptly because affected infants can undergo a precipitous deterioration in clinical status (14–16). Echocardiography allows rapid preoperative diagnosis with minimal risk to the patient, and eliminates the need for cardiac catheterization in most cases. Recently, Lincoln et al. (6) reported decreased mortality in patients who underwent repair of totally anomalous pulmonary venous connection without preoperative cardiac catheterization. A similar tendency was observed in our series, but this difference did not achieve statistical significance.

Postoperative evaluation. Postoperatively, echocardiography appears to provide an excellent means of assessing repair. In asymptomatic patients, if the Doppler recording from the individual pulmonary veins and the venoatrial anastomosis is phasic with a low peak velocity, if the Doppler color flow profile does not alias, and right ventricular pressure appears low by ventricular septal position or peak velocity of the tricuspid regurgitation jet, then routine postoperative cardiac catheterization should not be necessary. In patients with symptoms of pulmonary venous obstruction, the location and degree of obstruction can usually be determined with use of echocardiographic and Doppler techniques alone. Cardiac catheterization should be reserved for those patients whose site of obstruction cannot be identified, whose echocardiographic findings are inconsistent with the clinical course or in whom therapeutic catheterization (i.e., vascular stent placement) is contemplated.

Anastomosis size. Measurement of the cross-sectional area of the anastomosis provides a means of assessing surgical repair quantitatively. Intraoperative echocardiographic measurement of anastomosis size is feasible and could provide an immediate assessment of the adequacy of the surgical anastomosis. Serial postoperative measurements may be useful for long-term follow-up study of individual patients.

Transesophageal echocardiography. The relatively posterior location of the pulmonary veins makes transesophageal examination of pulmonary venous anatomy particularly helpful in patients with limited transthoracic echocardiographic windows. We have found transesophageal echocardiography to be an accurate means of examining pulmonary venous anatomy and obstruction in patients with complex heart disease (such as heterotaxy syndrome). The role of transesophageal echocardiography in the diagnosis and management of totally anomalous pulmonary venous connection in infants remains to be seen; however, it will undoubtedly facilitate the long-term follow-up of older children and adults after surgical repair of this lesion.

Conclusions. In this relatively small group of patients, our experience suggests that it is safe to repair totally anomalous pulmonary venous connection of all types without cardiac catheterization, when the diagnosis can be made satisfactorily by echocardiography. In addition, echocardiography appears to provide an excellent noninvasive means of evaluating patients after repair of totally anomalous pulmonary venous connection.

References

1. Smallhorn JF, Sutherland GR, Tommasini G, Hunter S, Anderson RH, Macartney FJ. Assessment of total anomalous pulmonary venous connection by two-dimensional echocardiography. Br Heart J 1981;46:613–23.
2. Snider AR, Silverman NH, Turley K, Ebert PA. Evaluation of infradiaphragmatic total anomalous pulmonary venous connection by two-dimensional echocardiography. Circulation 1982;66:1129–32.
3. Stark J, Smallhorn J, Huhta J, et al. Surgery for congenital heart defects diagnosed with cross-sectional echocardiography. Circulation 1983; 68(suppl II):II-129–38.
4. Huhta JC, Gutgesell HP, Nihill MR. Cross sectional echocardiographic diagnosis of total anomalous pulmonary venous connection. Br Heart J 1985;53:525–34.
5. Chin AJ, Sanders SP, Sherman F, Lang P, Norwood WI, Castaneda AR. Accuracy of subcostal two-dimensional echocardiography in prospective diagnosis of total anomalous pulmonary venous connection. Am Heart J 1987;113:1153–9.
6. Lincoln CR, Rigby ML, Mercanti C, et al. Surgical risk factors in total anomalous pulmonary venous connection. Am J Cardiol 1988;61:608–11.
7. Krabill KA, Ring WS, Foker JE, et al. Echocardiographic versus cardiac catheterization diagnosis of infants with congenital heart disease requiring cardiac surgery. Am J Cardiol 1985;56:351–4.
8. Cohn HE, Freed MD, Hellenbrand WF, Fyler DC. Complications and mortality associated with cardiac catheterization in infants under one year: a prospective study. Pediatr Cardiol 1985;6:123–31.
9. Sahn DJ, Allen HD, Lange LW, Goldberg SJ. Cross-sectional echocardiographic diagnosis of the sites of total anomalous pulmonary venous drainage. Circulation 1979;60:1317–25.
10. Lipshultz SE, Sanders SP, Mayer JE, Colan SD, Lock JE. Are routine preoperative cardiac catheterization and angiography necessary before repair of ostium primum atrial septal defect? J Am Coll Cardiol 1988;11: 373–8.
11. Freed MD, Nadas AS, Norwood WI, Castaneda AR. Is routine preoper-

ative cardiac catherization necessary before repair of secundum and sinus venosus atrial septal defects? J Am Coll Cardiol 1984;4:333–6.

12. Macartney FJ. Cross sectional echocardiographic diagnosis of congenital heart disease in infants. Br Heart J 1983;50:501–5.
13. Rice MJ, Seward JB, Hagler DJ, et al. Impact of 2-dimensional echocardiography on the management of distressed newborns in whom cardiac disease is suspected. Am J Cardiol 1983;51:288–92.
14. Clarke DR, Stark J, De Leval M, Pincott JR, Taylor JFN. Total anomalous pulmonary venous drainage in infancy. Br Heart J 1977;39:436–44.
15. Dickinson DF, Parimshahagan KM, Tweedie MCK, et al. Total anomalous pulmonary venous connection repair using deep hypothermia and circulatory arrest in 44 consecutive infants. Br Heart J 1982;48:249–54.
16. Turley K, Tucker WY, Ullyot DJ, Ebert PA. Total anomalous pulmonary venous connection in infancy: influence of age and type of lesion. Am J Cardiol 1980;45:92–7.